Case Letter

Spinular follicular keratosis of Siemens: A triad not to be ignored

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Sir,

Spinular follicular keratosis of Siemens (SFKS) is a rare X-linked or sporadic genodermatosis characterized by follicular hyperkeratosis and scar alopecia, palmoplantar hyperkeratosis (PPK), photophobia, corneal abnormalities, and atopy. Males are severely affected, yet females may also develop severe lesions in the context of usually X-linked inherited diseases. Herein, we report three cases of SFKS confirmed by biopsy.

The first case was a five-year-old boy from non-blood parents with a reported absence of hair at birth and no complaints of photophobia or atopy. No other similar cases were reported in the family. A dermatological examination revealed diffuse cicatricial alopecia of the scalp, eyelashes, and eyebrows and hyperkeratotic papules on the forehead and body associated with localized PPK and pachyonychia (Figs. 1a and 1b),

The second case was an eight-year-old boy from a nonconsanguineous marriage who had diffuse hair loss with depilation of the eyelashes and eyebrows since the age of two months, accompanied by photophobia, with no other similar cases in the family. A dermatological examination revealed scar alopecia on the scalp, complete depilation of the eyelashes, eyebrows, and body and follicular (Fig. 2) papules on the forehead and temporal region with localized KPP.

The third was a five-year-old girl from consanguineous second-degree parents with hair breakage and thinning

Figure 1: a) Palmoplantar keratoderma. b) Pachyonychia.

Figure 2: Frontal parietal alopecia with keratosis pilaris of the forehead.

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Figure 3: Woolly thinning hair.

without photophobia from three months of age, with a similar case in her younger sister. A dermatologic examination of both sisters revealed short, thin, woolly hair (Fig. 3), depilation of the eyelashes, eyebrows, and the rest of the body with the appearance of goosebumps, and plantar hyperkeratosis located on the heels of both feet.

A histopathologic examination of the scalp confirmed the diagnosis of SFKS in three of our patients, and an ophthalmologic examination revealed no abnormalities. All patients were prescribed topical keratolytic and emollient creams, and two of them were prescribed oral acitretin, which improved keratosis pilaris and palmoplantar without affecting the hair.

SFKS is a rare disease with X-linked or sporadic genetic transmission characterized by follicular hyperkeratosis and scar alopecia [1]. The pathophysiology of hair follicle destruction is not yet fully understood [2]. The disease usually begins in early childhood and worsens in adolescence, initially on the face progressing to the trunk and extremities [1]. PPK, photophobia, corneal abnormalities, and atopy may occur [3]. Therapy is not especially effective, and treatment is ineffective in the presence of changes that are mostly scarring. Although rare, SFKS should always be considered in all cases of follicular and/or palmoplantar hyperkeratosis with alopecia because, in addition to the genetic counseling necessary in some cases, treatment for this condition should be initiated as early as possible to delay and minimize scarring [2].

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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